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Tetralogy of Fallot and HIV infection in pregnancy: A case report

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Abstract

Tetralogy of Fallot (TOF) is the commonest congenital heart abnormality beyond the age of one year. Cases of Tetralogy of Fallot can present during pregnancy following successful surgical repair in childhood or occasionally as uncorrected TOF presenting for the first time during pregnancy, indeed the most frequently encountered congenital cyanotic heart lesion during pregnancy is cyanotic TOF. Most adult congenital heart disease in Africa is uncorrected due to widespread unavailability of cardiothoracic surgical services on the continent. Pregnancy is associated with significant haemodynamic alterations affecting both the systemic and pulmonary circulatory beds. These are more likely to have deleterious effects in pregnant women with underlying heart disease or with risk factors for pulmonary hypertension.

We describe here the case of a 22 year old pregnant woman with acyanotic tetralogy of Fallot and HIV infection who had an uncomplicated parturition. We discuss possible reasons why this potentially fatal combination was well tolerated by the patient.

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Introduction

Tetralogy of Fallot (TOF) is the commonest congenital heart abnormality beyond the age of one year. Cases of Tetralogy of Fallot can present during pregnancy following successful surgical repair in childhood or occasionally as uncorrected TOF presenting for the first time during pregnancy, indeed the most frequently encountered congenital cyanotic heart lesion during pregnancy is cyanotic TOF. Tetralogy of Fallot is a congenital cardiac malformation caused by a single developmental defect: an abnormal anterior and cephalad displacement of the infundibular portion of the interventricular septum. It consists of four components; an outlet Ventricular Septal Defect (VSD), obstruction to right ventricular outflow, an

overriding aorta and right ventricular hypertrophy, this may be associated with other congenital lesions such as Atrial Septal Defect (ASD). Most adult congenital heart disease in Africa is uncorrected due to widespread unavailability of cardiothoracic surgical services on the continent. Pregnancy is associated with significant haemodynamic alterations affecting both the systemic and pulmonary circulatory beds. These are more likely to have deleterious effects in pregnant women with underlying heart disease or with risk factors for pulmonary hypertension. The true incidence of Tetralogy of Fallot during pregnancy is difficult to ascertain, its contribution to morbidity and mortality is significant, accounting for up to 10% maternal mortality¹ and significant foetal morbidity in the western world.

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We describe here the case of a 22 year old pregnant woman with acyanotic tetralogy of Fallot and HIV infection who had an uncomplicated parturition. According to the Zimbabwe Ministry of Health And Child Welfare statistics, the prevalence of HIV infection among adults was 13.1% as of year end 2011, while the incidence rate is 0.85%. Data from antenatal surveillance is used to estimate and track the HIV epidemic in Zimbabwe, so these figures likely mirror the data among pregnant women in Zimbabwe. We discuss possible reasons why this potentially fatal combination was well tolerated by the patient.

Case Report

A 22 year old pregnant woman was referred to our hospital from a peripheral clinic for evaluation of a cardiac murmur. She was asymptomatic at the time. She was in the third trimester of her first pregnancy, and was seen for routine antenatal care by her primary care physician a week earlier. A murmur was detected on clinical evaluation and the patient was eventually referred for evaluation by the cardiologists at our institution. She had no symptoms suggestive of cardiac failure. She had been told as a young girl that she had a heart defect, but she was not aware of the actual diagnosis. She had been treated for heart failure before with a diuretic and potassium supplementation, but she had not needed this treatment for some years. She had no history of palpitations or documented arrhythmias, syncope or presyncope. She was HIV positive and was on zidovudine for prevention of mother to child transmission of HIV. Her last CD4 count was not known. She had never been on highly active anti-retroviral treatment. She had no history of tuberculosis or any other opportunistic events.

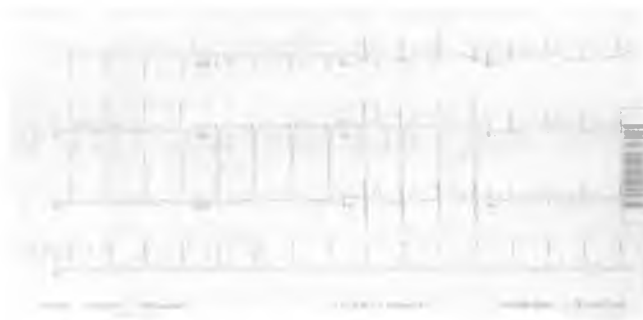
Examination revealed a well looking patient, exhibiting no signs of distress while at rest. She was not clubbed or cyanosed, and was pink. Her temperature was normal. She had no oedema. Her pulse was 96 beats per minute, regular and normal volume. Her blood pressure was 122/88 mmHg, her jugular venous pressure was 4 cm above the sternal angle, with normal waveform.

Her apex beat was not displaced, a left parasternal heave was appreciated and the pulmonary component of the second heart sound was palpable. Cardiac auscultation revealed a normal first heart sound and a loud second heart sound, she had no gallop rhythm. A pansystolic murmur was heard, grade 4/6 best heard at the lower left sternal edge with increased intensity during inspiration. A second murmur was heard at the second left intercostal space; ejection systolic murmur, and grade 3/6 intensity. These murmurs were consistent with tricuspid regurgitation and pulmonary stenosis respectively. Auscultation of her chest was normal. Her abdominal examination was normal. The obstetric examination showed a fundal height of 36 weeks, the foetus was in longitudinal orientation and cephalic presentation and the foetal heart sounds were

Normal.

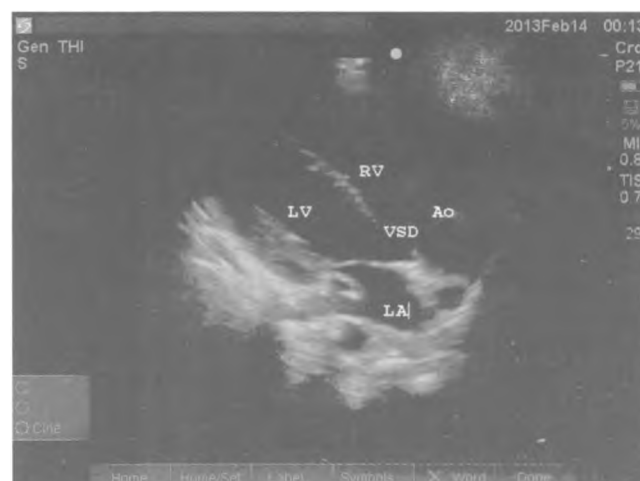
Her ECG showed regular sinus rhythm, with normal P waves and P-wave axis of approximately + 60 degrees, the PR interval was 160 ms. The QRS axis was approximately +150°, right axis deviation. There was evidence of right ventricular hypertrophy and inverted T waves in lead V1 (Figure I).

Figure I: Electrogram showing right axis deviation, right ventricular hypertrophy and inverted T waves in leads V1 and III.



Her chest radiograph showed mild cardiomegally and clear lung fields. Echocardiography showed the left ventricle to be normal in size and function, ejection fraction estimated at 60%. There was moderately severe left ventricular hypertrophy. The left atrium was mildly enlarged. The right ventricle was dilated, with poor systolic function. There was severe right ventricular hypertrophy. A small high septal ventricular septal defect was noted. The aortic root was mildly dilated and the aorta was overriding the right and the left ventricle. The pulmonary valve appeared hyperechoic and thickened, there was a pressure gradient of approximately 105 mmHg across the pulmonary valve. There was moderately severe tricuspid regurgitation. The mitral valve and tricuspid valve appeared normal. An echocardiographic image is shown in Figure II.

Figure II: An echocardiographic image of the patient. Parasternal long axis view, showing the Ventricular Septal Defect (VSD) and the aorta overriding the right and left ventricles.



Obstetric ultrasound showed a single viable foetus with no obvious congenital abnormalities, estimated gestational age was 36±3 weeks. FBC showed a mild normocytic anaemia; WCC 6.1, Hb 8.9 MCV 81.8 Platelets 205. CD4 count was 836 cells/μl. Renal function was normal.

A diagnosis of acyanotic Tetralogy of Fallot with pulmonary hypertension was made. HIV infection was an important co-morbidity. The patient was admitted into hospital under the care of obstetricians, physicians and anaesthetists. She was continued on zidovudine and delivered a normal foetus, birth weight 2800 grams after induction of labour. Delivery was at 37 weeks gestation. She was given prophylactic antibiotics at the onset of labour. The baby was given nevirapine at birth and was to continue on nevirapine for at least another six weeks as the mother was going to breast feed for that duration.

A repeat transthoracic echocardiography examination after delivery showed the same findings except reduced transpulmonic pressure gradient and downgrading of the severity of tricuspid regurgitation from moderately severe to mild. Unfortunately, actual pressures were not given at this time. The immediate post-partum period was uneventful and the mother and child were discharged home. The decision to deliver vaginally was taken by the multi-disciplinary team, in recognition of the high operative risk of patients with pulmonary hypertension and the need to minimize risk of transmission of HIV infection from the mother to the child. Since the mother had been on anti-retroviral therapy to reduce risk of transmission, the operative risk to the immediate well being of the mother was considered too high and hence the decision to deliver vaginally.

Discussion

Pregnancy is associated with significant haemodynamic changes which have potential to precipitate or worsen symptoms in women with pre-existing cardiac disease. The main changes are an increase in plasma volume and cardiac output of up to 50% peaking in the second trimester and a fall in both peripheral and pulmonary vascular resistance due to vasodilation.³ Congenital heart disease in pregnancy has been associated with a marked increase in risk of adverse cardiovascular events, especially supraventricular arrhythmias and death.⁴

Maternal and foetal outcomes in pregnancy complicated by congenital heart disease are predicted by history of a prior cardiac event, baseline functional status or presence of cyanosis, presence of left heart obstruction, reduced left ventricular systolic function.⁵ Maternal arterial oxygen saturation and haemoglobin level can be used to predict risk of adverse foetal outcome.⁶ For this reason a pregnant woman with congenital heart disease presents complex challenges for the obstetrician, physicians and anaesthesiologists managing the case and indeed, a multidisciplinary

effort is recommended to optimise outcomes.³

Pregnancy outcomes in women with tetralogy of Fallot have been described in the literature in case reports,^{1,7} and the fatal combination of pregnancy, HIV infection and HIV associated Pulmonary Arterial Hypertension (PAH) has been documented.⁸ The prevalence of HIV PAH in developed countries is estimated to be approx. 0.5% (similar to that in the pre HAART era).⁹ In the developing countries a higher prevalence is estimated; an echocardiographic study in West Africa's Burkina Faso estimated prevalence to be 5%.¹⁰

Pulmonary hypertension, especially if severe tends to be poorly tolerated during pregnancy, irrespective of the underlying aetiology.¹¹ The outcome for both mother and child was good in the case we have described, despite potentially fatal interactions between the congenital heart disease, HIV infection and the haemodynamic effects of pregnancy. The degree of cyanosis and haemoglobin level have been strongly linked to adverse maternal and foetal outcome.⁶ Our patient was not cyanosed throughout the course of her pregnancy and this would have favoured a good outcome. Despite this careful monitoring was still important because of potential for decompensation in late pregnancy or during parturition. The fall in total peripheral resistance and hypotension in parturition both increase the right to left shunt resulting in hypoxemia and emerging or worsening cyanosis.¹² A similar case was described in multiparous women who presented with cyanosis from an unrepaired TOF only in the third pregnancy.¹³

In the case presented the pressure gradient across the pulmonary valve of 105 mmHg would suggest a severely stenotic right ventricular outflow tract which would be unlikely to be well tolerated during pregnancy. This was probably a result of an apparent increase created by a fall in pulmonary vascular resistance associated with the pregnant state.³ Maternal haemoglobin of less than 18 g/dl has also been associated with a higher likelihood of favourable pregnancy outcome in women with cyanotic heart disease.⁶ Our patient falls into this category, and was actually anaemic, probably a combination of pregnancy related haemodilution, nutritional deficiencies and potentially HIV infection.

Our case highlights a unique combination of uncorrected acyanotic tetralogy of Fallot, HIV infection and pregnancy which were remarkably well tolerated by the patient who had a good pregnancy outcome. To the best of our knowledge, this has not been described before in the literature.

Cases of tetralogy of Fallot without cyanosis have been described in Africa before,¹⁴ however this was not in the setting of pregnancy or another risk factor for pulmonary hypertension such as HIV infection and no prior surgical correction of the congenital abnormalities.

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